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ON DESMOID TUMOURS AND THEIR AETIOLOGY.

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- ON DESMOID TUMOURS AND THEIR AETIOLOGY.

The term "Desmoid", given originally by Johannes Mueller to certain tumours of "tendon like consistency" arising in the abdominal wall, has been restricted by Pfeiffer (1) to fibromata or fibrosarcomata arising from the musculo aponeurotic structures of the abdominal parietes, and the use of the term thus is authorised in v. Bergmann's Handbuch de Praktischen Chirurgie.

In our own language the term "Desmoid" has been adopted by some authors and is frequently used clinically to designate this special type of tumour: it is so used in Keen's 'System of Surgery' and several cases have been reported under it.

As to my mind these tumours have special characteristics which distinguish them from the fibrosarcomata, I will continue to use the term, - as above defined by Pfeiffer, - throughout this thesis.

I have chosen this as my subject because as Resident Medical Officer to the Chelsea Hospital for Women, I have seen two cases during the past year; both of these cases were operated on and the tumour carefully examined.

Further, we have had two other examples of this variety of tumour in this hospital during the last 10 years, and of these cases also we have the clinical notes and pathological slides.

A still further reason for this choice is that little has been written in English on this subject, for a careful search through our medical literature of the last 20 years has resulted in my finding but two short papers on the subject.

One of these, read by Mr Alban Doran (2) before the London Medical Society, was based on a case which he had operated on, and was followed by a short discussion by the Society, in which Mr Bland Sutton said that he considered the use of the term "Desmoid" inadvisable, as, in his opinion, all these tumours were Sarcomata.

The second paper is one by Mr Lacy Firth (3); this paper also is based on a single case.

Note 1. - C. Pfeiffer: Beiträge Zur Klinischen Chirurgie 1904.
Die Desmoide der Bauchdecken und ihre Prognose.

Note 2. - Doran: - Fibrosarcoma or Desmoid growth of the abdominal wall. Med.Soc. Transact. xiv.

Note 3. - Lacy Firth: Bristol Med. Chir. Journal 1906.

Apart from these papers I have found only reports of single or short series of cases, amounting to about 20 in all.

That these characteristic tumours are rare is undoubtedly true, still we must conclude that many have been classed as ordinary fibromata or fibrosarcomata and not considered worthy of report, because as soon as we turn to the literature of other countries we find a much larger number of recorded cases.

Thus Pfeiffer in Germany was able to collect 400 cases, and his classical monograph, which appeared in 1904, contains a record of the great majority of the cases published before that year. His 400 cases are made up as follows :-

- 40 cases of his own which had been seen in v. Bruns clinic at Tübingen during a period of 46 years.

100 cases previously collected and published by Ledderhose.

260 cases culled from the literature.

His monograph is the outstanding work on the subject and I will refer frequently to his statistics and to the conclusions he formed from a consideration of this large number of cases.

Since 1904 the papers of note are :-

1. That of Lacy Firth previously mentioned.
2. An article by Harvey B. Stone, "Annals of Surgery" 1908.
3. Remarques sur les fibromes de la paroi abdominale - P. Legere et Delamare, "Le Progrès Medical" 1908.
4. A report of a clinical lecture on Tumours of the Abdominal Wall by Lockwood is published in the "Lancet", August 1910. Lockwood refers briefly to 7 cases, some of which appear to be of this variety.

This thesis of mine is based on a study of the above papers /

papers and monographs and I will compare their conclusions with those which I have formed from my own cases. I have in addition gone over all the cases recorded in the French Literature of the last 20 years and have found 8 cases which are not included in Pfeiffer's monograph.

I have referred above to the comparative rarity of these tumours. Pfeiffer quotes Guerlt as finding .13% of Desmoid tumours in a series of 16,630 tumour cases collected from the statistics of the Vienna Hospitals.

I will now proceed to give the Clinical Histories of my 4 cases.

Case 1. - G.F. - aged 26; 1 Child; no miscarriages. Admitted September 26th 1911 under Mr Victor Bonney. Patient was a robust, healthy-looking woman.

History. - About 8 months ago patient noticed accidentally a lump in the abdominal wall which felt "like a hardness in the muscle about the size of a penny". The lump lay immediately above and to the right of the umbilicus and gave rise to no symptoms.

Patient was confined 1 year and 11 months ago. The labour was non-instrumental and easy.

Patient at no time subsequent to labour has been conscious of any weakness or pain in the abdominal muscles. There is no history of trauma or abdominal strain.

Previous Health - has been excellent.

On examination - a rounded hard tumour about the size of the closed fist is felt immediately above and to the right of the umbilicus. The tumour appears to lie in the substance of the Right Rectus Muscle and is movable over the subjacent intra peritoneal structures.

Operation - A hard tumour, size of closed fist, was disclosed in the substance of the Right Rectus Muscle. The muscle fibres seemed to disappear abruptly into the tumour which was adherent to the anterior sheath of the Rectus. The tumour was cut out with free haemorrhage. The posterior sheath /

sheath of Rectus and peritoneum were found to be firmly adherent to its posterior aspect and portions of these structures had to be removed with the tumour. The gap left in the peritoneum and muscle was closed, with some difficulty, with strong silk mattress sutures.

Progress - Uneventful and union satisfactory.

After History - Patient was in good health and showed no sign of recurrence 5 months later.

Case 2. - B.M. - aged 34. Inpatient under Mr Giles,
7/7/1911 to 23/7/1911.
Four children, aged respectively 6 years;
4 years, 3 years, and 16 months. 1 miscarriage 8 years ago.

Patient had severe abdominal pains after this last confinement and then noticed that she could feel a lump in the lower part of the abdomen. She drew her Doctor's attention to it but he made light of it and told her it was only "after pains". Patient got about at usual time after this confinement and had no further trouble until quite recently when she again had occasion to consult her Doctor on account of increasing loss at the menstrual periods.

On examination the lump in hypogastrium was found and patient was sent in with the diagnosis of Fibroids of Uterus.

Previous Health - excellent.

Obstetrical History - 1st, 2nd and 3rd labours were rather prolonged and severe; 1st was instrumental; the last (4th) was an easy labour.

Patient was a strong, healthy-looking woman.

On abdominal examination an ill-defined tumour was felt above the pubis and to left of middle line.

P.V. The uterus was bulky and bimanually the lump in /

in hypogastrium seemed to be connected with the uterus. A diagnosis of subperitoneal fibromyoma of uterus was made.

Operation: - A median suprapubic incision was made, when the entire tumour was found to lie in the back of the lower end of the left Rectus muscle. the tumour lay embedded in the muscle and measured about $1\frac{1}{2}$ by 2 inches, with its longer axis vertical.

All the deeper and outer portion of the muscle fibres seemed to terminate abruptly in its wall. The tumour had to be cut out of the muscular substance and it was found to be firmly adherent to the peritoneum which was opened in removing it.

The tumour, which was very hard and well-defined, was intimately adherent to the muscle on all its aspects except posteriorly where the peritoneum covered and adhered to it.

The uterus was found to be bulky and sub-involuted but otherwise healthy.

The gap was drawn together with strong silk mattress sutures and the wound healed satisfactorily.

After History: - Seven months later patient was found to be in perfect health and showed no sign of recurrence.

Case 3. - E.G. - aged 35. Inpatient 26/2/1900 to 20/3/1900 under Mr Bland Sutton.

Obstetrical History -

1st January 1890, - non-instrumental and easy.
2nd April 1895 , - very difficult and prolonged.
3rd Sept 1897 , - difficult.

Patient states that the 2nd confinement left a weakness in the right inguinal region, which was noticed when she wiped her foot on mat or trod heavily on anything. This feeling of weakness became more marked and patient had pain in the same /

same situation off and on during the last 5 months of the 3rd pregnancy.

The presence of a tumour was noticed about six months after this 3rd confinement, and has grown steadily but practically painlessly up to date of admission.

Examination: A solid rounded mass is situated in the Right Inguinal Region; it appears to be partly fixed, is dull on percussion, and about the size of a large orange.

Operation: An oblique incision was made over the tumour, and the tissues cleared from its anterior and lateral aspects. Posteriorly the tumour was found to be firmly adherent to the fascia of the internal oblique. The entire tumour lay firmly embedded in the substance of the abdominal wall.

Haemorrhage was not excessive during removal, but several fragments of new growth were left, and had to be dissected away after removal of the main tumour. The peritoneum was laid bare but not injured. The wound was closed with strong silk and silk-worm gut.

Progress - was uneventful and union satisfactory.

Pathological Report - Fibroma.

This patient in December 1911 reported herself as well and with no sign of recurrence.

Case 4. - E.B.S. - aged 24; Inpatient March 13th - March 27th 1903 under Mr Bland Sutton.

Patient first noticed a small lump, size of walnut, in Right side of lower abdomen 14 months ago. It gave rise to no pain and grew but slowly. One month ago patient was confined and during parturition the lump increased rapidly in size.

2 children; no miscarriages. (The age of oldest child is unfortunately not stated).

Examination: /

Examination: In right Iliac Region there is a swelling about the size of a very large orange which seems to be situated in the substance of the abdominal wall. It is of firm consistence and seems to be connected with the Iliac bone. It is painless and slightly movable. There is no fixation of skin over it and no increase of vascularity around it.

Operation: An incision was made down to the tumour almost parallel with but above Poupart's ligament. The tumour appeared to arise from and was firmly adherent to the musculo-aponeurotic structures of the abdominal wall. Posteriorly the peritoneum stripped off without injury.

Progress - was satisfactory.

Pathological Report - Fibroma.

December 1911. The address of this patient is now unknown.

Let us note with regard to these four cases :-

1. That all were in women.
2. That all were in women who had borne children and were within the child-bearing age.

Of Pfeiffer's 400 cases -

87.1%..... were in women and of these women
94.3%..... had borne children.

Let us now examine the cases recorded since the publication of Pfeiffer's monograph with regard to these and other essential points. -

Harvey B. Stone's paper ("Annals of Surgery" 1908) includes the following with references to place of publication:-

1. Cullen /

1. Cullen - Mrs N.M., aged 30 years; no note of pregnancies. Tumour in left hypochondrium - lobulated and freely movable attached to muscles.
- Pathological Report - Pure Fibroma.
2. Schwarzschild - Woman, aged 28 years.
Tumour Right side of abdomen. Rapid growth during pregnancy.
- Pathological Report - Fibroma.

One year later there was no recurrence.
3. -do- - Woman, aged 30 years; 2 children.
Tumour appeared to spring from post sheath of Rectus and occupied the Right lower quadrant of that muscle.
4. Eitel - Mrs J. aged 26 years.
No note as to pregnancies.
The tumour was situated above and to left of symphysis pubis and was deeply embedded in the abdominal wall.
- Pathological Report - Very cellular Fibroma.
5. Gross & Sencert. - Woman.
Tumour 6 kilos in weight; 10 years duration. The tumour was attached by broad pedicle to anterior sheath of Rectus. Skin was ulcerated over the projecting tumour.
- Path. Report - Gangrenous lipo-Fibroma.
6. McAdam Eccles (4) - Woman, aged 37 years; 2 children.
Elongated swelling in left Rectus muscle running from inch above umbilicus to half-way between umbilicus and pubis. The tumour appeared to grow from the front of Rectus sheath and to infiltrate the muscle.
- Path. Report - Fibro sarcoma.

18 months later there was no recurrence.

1. /

7. McAdam Eccles. - Single woman, aged 26 years.
Tumour in abdominal wall of Right Inguinal Region, - appeared to spring from the Internal Oblique near Poupart's ligament.
8. -do- - Woman, aged 35 years.
3 children, youngest 3 years.
Tumour of about 6 months duration, situated just internal to left Anterior Superior Iliac Spine. The tumour appeared to be attached to peritoneum.
9. Bidwell (5) - Woman, aged 36 years.
Tumour in Right Inguinal Region of 18 months' duration. Patient was confined 1 year ago. The tumour lay in the fibres of the Transversalis Muscle and occupied the Right Inguinal Canal.
- Path. Report - Fibro Sarcoma.
10 years later no recurrence.
10. -do- loc.cit. Mrs H. aged 33 years.
5 children; 1 miscarriage 6 months ago, and now 2 months pregnant.
Tumour in upper part of Right Rectus Muscle which appeared to spring from the posterior sheath of Rectus.
- (Bidwell remarks that in both his cases the tumours seemed to bear some relation to pregnancy).

HARVEY STONE found 5 cases of Tumour of Abdominal Wall in the records of Halstead & Bloodgood's Clinic. The first case he himself excludes as being probably a subcutaneous nodule. The other four are as follows :-

11. - Woman, aged 56 years; 1 Child 21 years of age.
The tumour appeared shortly after this confinement and was operated on a year later. The growth slowly recurred in the original site and /

and was removed at a second operation 20 years after the first.

- Pathological Report - Spindle called Fibroma.

12. - Woman, aged 23 years. Tumour followed pregnancy. Tumour was situated below and to left of umbilicus and was attached to posterior sheath of Rectus. The growth infiltrated the deeper structures of the abdominal wall.

13. - Woman, aged 35 years. Rapidly growing tumour of Right lower quadrant of abdomen following pregnancy. Part of Rectus muscle was involved and removed with the tumour.

14. - Coloured Woman, aged 18 years. - Tumour appeared during pregnancy to left of and just below the umbilicus. It was adherent to the anterior sheath and infiltrated the Rectus muscle.
- Path.Report - Spindle called sarcoma.

(Stone remarks that this is the only recorded case of Desmoid tumour in a coloured woman).

Stone himself records the following case :-

15. - Mrs A.G. aged 23 years; One child still-born; instrumental labour (date not stated). Tumour was noticed 7 months ago. 2 cm. to left of umbilicus, there is an ill-defined ovoid mass 8 x 5 cm., elongated in the long axis of the Rectus muscle. The tumour was thought to be intra-abdominal.

At operation the tumour was found to be situated entirely within the abdominal parietes. It was adherent to and appeared to arise from the posterior sheath of the Rectus muscle. The tumour involved the entire thickness of the muscle in the median two-thirds of its width. The tumour projected back into the abdominal cavity and this aspect of the tumour was firmly adherent to the peritoneum.

- Path.Report - Very cellular Fibroma.

LOCKWOOD in Lecture on "Fibroma, Sarcoma and Fibromyoma of the Abdominal Wall" ("Lancet" August 13th 1910) gives 4 cases of Fibroma, 2 of Fibrosarcoma and 1 of Fibromyoma,

One of his cases of Fibroma was in a Man, aged 32.
Tumour was of 14 years duration. At operation the tumour was found to lie between the Rectus Abdominis Muscle and its posterior sheath and was easily shelled out.

(We consider that this tumour was not a Desmoid, for reasons which we put forward later).

Lockwood's other six cases were in Women, but no note is given as to presence or absence of history of pregnancy in any of them :-

16. - Jewess, aged 15 years.
Tumour of anterior sheath of Rectus; two years duration.
- Path.Report - Fibroma.
17. - Woman, aged 25 years.
Tumour of left conjoint Tendon.
- Path.report - Fibroma.
18. - Woman, aged 24 years.
Tumour situated in Right Semilunar line near McBurney's point.
- Path.report - Fibroma or Fibro-sarcoma.
Tumour was operated on, on a diagnosis of appendicular abscess.
19. - Woman, aged 30 years.
Patient had, what was diagnosed as a Fibroma, removed from amidst the fibres of the external Oblique muscle at level of the 11th Costal Cartilage. 22 months later she had a recurrence at same level and tumour was adherent to the front of the Rectus Sheath.
- Path.report of the second tumour, - Spindle celled Sarcoma.
After History - unknown.

20. - Woman, aged 20 years.
Patient had a blow on abdomen in 1904. In 1906 a tumour appeared in the Right Semilunar Line, about the level of the umbilicus. This tumour was removed abroad and recurred 9 months later.
In Sept 1908 recurrence in scar was widely removed.
In December 1909 Nodules recurred in scar.
In July 1910.....Fourth operation for further local recurrence.
- Path.report - Spindle celled sarcoma.

Lockwood's seventh case appears to have been an instance of Fibromyoma arising from the Round Ligament of the Uterus.

21. - Woman, aged 33 years.
Presence of tumour was noticed in November 1908. At operation in January 1909 an oval tumour was found stretching from Anterior Superior Iliac Spine to Crest of Pubis.
- Path.report - Fibromyoma.

22. - (Lacy Firth's case) - Woman, aged 29 years;
3 children, youngest 2½ years.
Swelling noticed in left Iliac Region for 4 months. Pain in left groin on exertion for 2 years.
At operation the inner portion of tumour was found to blend with the Internal Oblique and Transversalis muscles. The main bulk of the tumour lay comparatively free in the sub-peritoneal fat.
- Path.report - Fibro cellular or connective tissue tumour.

23. - (Krusen's case (6)) - Mrs J.H. aged 29 years.
4 children; eldest 9 years; youngest 11 months.
Tumour of abdominal parietes of one year's duration. At operation a hard well-defined tumour was dissected out of the substance of lower quadrant of left Rectus muscle.
- Path.report - Fibroma.

The /

The following cases are not recorded in Pfeiffer's Monograph, although they appeared previous to the date of its publication :-

24. - Briddon - (New York Medical Record, March 1882).

Woman, aged 27 years; 3 children.

Tumour was first noticed during the second month of the last pregnancy. It was situated in Right Iliac fossa and measured 9 x 5 inches. Tumour at operation was found to be incorporated with the fibres of the Transversalis and Int. Oblique Muscles.

- Path.report - Fibroma.

25. - Wallace - (Liverpool Med. Chir. Journal, 81-82).

Woman, aged 34 years; 5 children; youngest aged 4 months.

Patient noticed the presence of a small lump below and to the left of the umbilicus before the commencement of her 4th pregnancy.

At operation a tumour, size of cocoanut, was found in the Rectus muscle and it appeared to take origin from the posterior sheath of Rectus to which it was firmly adherent.

- Path.report - Fibro sarcoma.

26. - Fournell - (Congrès Français de Chirurgie - 1891)

Woman, aged 27 years; 4 children; youngest 2 years.

Eight months after this last confinement patient got a knock on the abdomen and found on examination that there was a lump, size of nut, between the umbilicus and Iliac Spine.

At operation tumour was intimately adherent to the Oblique&Transversalis muscles with the peritoneum adherent to it posteriorly.

- Path.report - Fibroma.

27. - Silberberg. - (Bulletins de la Soc. Anatomique - 1903)

Mrs P.I. aged 23 years; 3 children, youngest aged 4 years.

Tumour in Right Inguinal Region of 10 months' duration /

duration. At operation the muscles of the abdominal wall appeared to run into the tumour.

- Path.report - Fibroma with cellular areas.

28. - Abadie. (Bulletins de la Soc. Anatomique - 1903).

Mrs L.M. aged 30 years; 4 children -

9; 7; 3 years, and 5 months.

Tumour in abdominal parietes of left Iliac Region. Development of tumour appeared to be associated with this last pregnancy.

29. - P.Fredet. (Bulletins de la Soc. Anatomique - 1903).

i. Woman, aged 31 years; 3 children.
Tumour in substance of Right Rectus immediately above umbilicus.

30. - ii. Woman, aged 24 years; 3 children.
Tumour in left Ilio Inguinal Region was noticed $2\frac{1}{2}$ years ago during course of second pregnancy. Since then patient has taken a third child to term. At operation the tumour was found to be incorporated with the Oblique Muscles.

- Path.Report - Fibroma.

The edge of section shows muscle fibres gradually merging with the tumour substance.

31. - Vernot. (Journal de Medecine de Bordeaux - 1903).

A.C. aged 21 years; 2 children; 3 and 1 years.

Patient gave history of slight trauma.

The tumour was incorporated with the muscles of the left Ilio Inguinal Region.

(Vernot thinks the tumour was due either to accidental trauma or to trauma associated with labour).

- Path.report - Connective tissue tumour.

Lockwood's case of Fibroma of abdominal wall of 14 years' duration in Man of 32 years is excluded from the above series because it is described as having been shelled out from the cellular tissue behind the Rectus Abdominis Muscle. It therefore fails to come under our original definition, which requires /

requires that the tumour must be connected with the musculo aponeurotic structures of the parietes.

The fact that my own four cases were all in women means little, because they occurred in the practice of a Hospital for Diseases of Women; but the 31 additional cases given above were practically all described in general medical literature, and the only selection exercised in their choice is that they must satisfy our original definition.

On the strength of these 35 cases, and for certain other reasons which I will put forward later, I am inclined to go further than Pfeiffer (who found 87% in women) and I would question whether a true Desmoid tumour ever occurs except in women.

Again, of my total of 35 cases, the fact that there has been antecedent pregnancy is recorded in 26. In the remaining 9 (Lockwood's 6 cases and 3 of other authors) the presence or absence of this important factor has not been noted in the history of the case. Pfeiffer states that of his cases occurring in women 94.3% of these women had borne children: and on this point also my figures would give quite as high a proportion. This fact adds another point of interest in practically limiting the occurrence of Desmoid tumours to the child-bearing age: for in the one or two exceptions recorded, the tumour has been of many years' duration as evidenced by the history and by its great size.

Having now considered Desmoid tumours as regards frequency, and their incidence as to sex, age, and antecedent pregnancy, we will now pass on to enquire their site, and the structure from which they take origin.

Of our 35 cases -

18, or just over 50% were described as involving the Rectus muscle or the Rectus muscle and sheath, and most of them were situated below the umbilicus.

40% in the Illo Inguinal Region in nearly equal proportion - Right and Left.

10% as arising in the abdominal wall at some other point.

Pfeiffer /

Pfeiffer found :-

43% in Rectus muscle or sheath.
17.5% in other abdominal muscles.
17% in superficial or deep abdominal fascia.

and adds that in many cases several muscles or fascial layers were involved so that the original point of origin was not easily discoverable.

In general terms, of his 400 cases, he found that 72% were situated below the umbilicus.

We may therefore conclude that Desmoid tumours are found in the majority of cases to occupy the lower half of the abdominal wall, and to take origin in the Rectus Muscle and sheath or in the musculo aponeurotic structures of the Ilio Inguinal Regions, but that they may occasionally be found at any point of the abdominal parietes.

As to the structure in which they arise, older writers on the subject invariably stated that this appeared to be an aponeurotic or fascial structure and this assertion is frequently repeated in the reports of more recent cases.

A study of our two tumours undoubtedly negatives any such conclusion.

In both our cases the tumour arose in the substance of the Rectus Muscle, and where the growing edge of the tumour reached the sheath, it adhered to it and to the subjacent peritoneum. The tumour then showed no further tendency to perforate or permeate those adherent fascial structures; and I am inclined to think that such limitation is characteristic, because of the fact that in no case has an intraperitoneal spread ever been recorded.

- AETIOLOGY. -

We now come to consider the Aetiology of Desmoid tumours. Much has been written on this and many theories put forward by earlier writers now stand disproved.

Thus Nélaton in 1862 (7), in describing a tumour which was apparently of this nature, pointed out that the tumour appeared /

appeared to be connected with the Iliac bone by a well-marked fibrous pedicle, and stated that in his opinion these tumours were of periosteal origin. The fact alone that Desmoid tumours are most commonly situated in the Rectus Muscle, and that no evidence whatever of any connection with bone or periosteum can be found on removing them is sufficient to throw Nélaton's theory out of court.

On similar ground we would discard Guinard's (8) theory which is, - that these tumours take origin in the Round Ligaments, for the great majority of these tumours are situated remote from, and quite unconnected with, these structures.

Hertzog (9) has suggested, as a more likely explanation of their origin, that trauma of the abdominal muscles might give rise to a haematoma which gradually formed a keloid cicatrix and later took on fibro-sarcomatous characteristics which resulted in tumour formation.

Against this theory we have several facts :-

- i. There is no history of direct trauma in the great majority of cases.
- ii. Haematomata of the abdominal muscles resulting from injury are of very great rarity and where they have occurred they have followed the usual course of haematomata of muscle elsewhere.
- iii. As regards the possibility that these tumours might be analogous to keloid, Stone loc.cit. notes in recording a case in a negress that this is the only case recorded in a coloured woman, whereas the dark-skinned races show a much greater liability to develop keloid in cicatrices elsewhere.
- iv. If Desmoid tumours were of traumatic origin, we would expect to find a much larger proportion in men because of their much greater exposure to injury.

For /

Note 8: - Guinard - In Traité de Chirurgie par Le Dentu
et Delbet - Fibromes des parois Abdom.

Note 9: - Ueber Fibrome der Bauchdecken - München 1883.
(Vi. Hl 83 ii p. 414).

For the foregoing reasons I think we may discard the purely traumatic theory of origin and we will pass on to consider Pfeiffer's Theory as to their Aetiology.

Pfeiffer points out that Wischnewsky has shown in certain researches that there is in pregnancy - hypertrophy of the abdominal muscles coincident with the hypertrophy of the uterus, and strikingly different to the atrophy of the abdominal musculature, which is found in all pathological causes of abdominal distension, eg. as in ascites, ovarian cyst, etc., and he considers that Ribbert's Theory as to tumour origin might hold good in the case of Desmoid tumours, i.e. -

That here we have a tissue hypertrophy plus an alteration in the tissue tension which would be sufficient to account for the occurrence of occasional persistent and progressive hypertrophy of isolated areas of muscle - the actual site of such hypertrophy being perhaps determined by accidental trauma or in injury sustained during labour.

Now, it is not surprising to my mind that we should find a striking difference between the muscular tissue of the abdominal wall in a patient suffering from ascites and general malnutrition when contrasted with the abdominal muscles of a woman in the physiological condition of pregnancy; but that there is any actual hypertrophy of the muscles of the abdomen in pregnancy is not by any means proved.

Dr Eden states that there is actual mechanical thinning of the abdominal wall in the later stages of pregnancy and that this muscular thinning is always strikingly noticeable during the performance of Caesarean Section.

We may therefore safely conclude that the persistence of theoretical hypertrophy in an isolated area of muscle is still more difficult to prove and we will pass on to search for a more satisfactory explanation to account for these tumours.

Let us remind ourselves at the outset that every tumour in the abdominal wall is not necessarily a Desmoid tumour.

Pure Fibromata, Fibro-sarcomata, Fibro-myomata, and Sarcomata occur here as elsewhere in the body. What then are the characteristics /

characteristics of a true Desmoid tumour.?

The very briefest study of the recorded cases will show us that two factors are practically constant in all of them.

1st - That these tumours are almost entirely confined to women.

2nd - To women who have borne one or more children.

Pfeiffer notes in his monograph that the tumours classed as "Desmoids", which were found in the male sex, differed histologically from the apparently similar tumours occurring in the female, in that they were more cellular, more resembling fibro-sarcoma in type, and further he notes the significant fact that the prognosis as regards malignancy, judged by recurrence after operation, was worse in men. His figures are as follows :-

22 men	Recurrence 68.1%Mortality 50%.
85 women.....	Recurrence 21.2%Mortality 7%.

Those statistics show clearly in my opinion that the tumours described as "Desmoid" and as occurring in the male sex were not true Desmoids, but probably of a sarcomatous nature.

Thirdly, I would state here, that a true Desmoid tumour has certain histological characteristics which distinguish it from all these other varieties of tumour growth, which may also occur in the abdominal wall and hence be confounded with it.

These characteristic features of the Desmoid tumours are seen when sections are studied, cut from the growing edge of the tumour, where the tumour substance merges with and infiltrates the muscle in which it grows.

And I am led to believe from the fact that certain characteristic cellular elements are constantly found in this growing edge, and from the fact that antecedent pregnancy is an almost constant factor in the previous history of all the authentic cases recorded, that these tumours are intimately connected with and owe their origin directly to this pregnancy.

And further, this association with pregnancy is not an accidental one, - not that theory of many writers who noticed the great frequency of preceeding pregnancy and ascribed the origin /

origin of these tumours to an accidental rupture of muscle fibres during labour.

This theory is aptly stated by a French writer (10) as "Petit rupture musculaire, foyer hemorrhagique, proliferation fibreuse", but fails absolutely to supply an adequate reason for the origin of these tumours, because, as we have before stated, traumatic ruptures and injuries of muscle or fascia never go on to tumour growth, but in all cases end in simple fibrous cicatrices.

We must look more deeply for an explanation.

We grant that it is probable that a localised muscular injury determines the actual site of tumour growth, - but what causes it.?

And the appearance of the characteristic cellular elements which are invariably found in the tumour edge suggests an explanation which, to my mind, has more to support it than any other theory previously suggested.

For in all sections through the growing edge we find certain protoplasmic masses which lie grouped about the broken up and degenerating ends of the muscle fibres, and these cellular elements bear a striking resemblance to the syncytial masses which are characteristic of the primary and secondary growths of Chorion-epithelioma Malignum - (v. fig. 1.).

Now, that multinuclear cell masses occur in the tumour edge is not a new observation: these cell masses have been noticed and commented on in several cases recorded by French Surgeons. Thus, Legène and Delamare in "Le Progrès Medical" 1904 wrote as follows :- "We take exception to any account of these tumours which describes them as encapsulated; the muscle is infiltrated where it merges with the tumour, and at certain points we have observed the curious alterations of retrogression of the striped muscle tissue which Durante has so well described under the name of false giant cells. The origin of these giant cells is apparent because they show the staining reactions of sarcoplasm".

In their opinion, therefore, these giant cells are derived from the degeneration and breaking up of the invaded muscle fibres.

The /

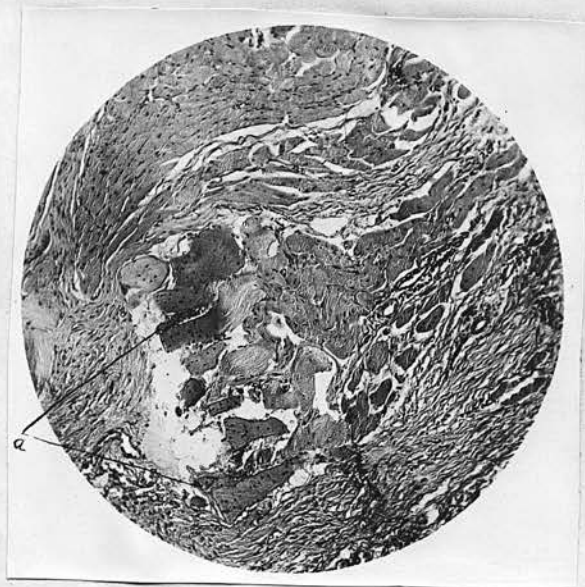


Fig. 1. - Microphotograph of section, Case 2, B.M.

$\frac{2}{3}$ objective; No.5 eyepiece.

Note the multinucleated protoplasmic masses-a
lying in contact with the degenerating muscle.

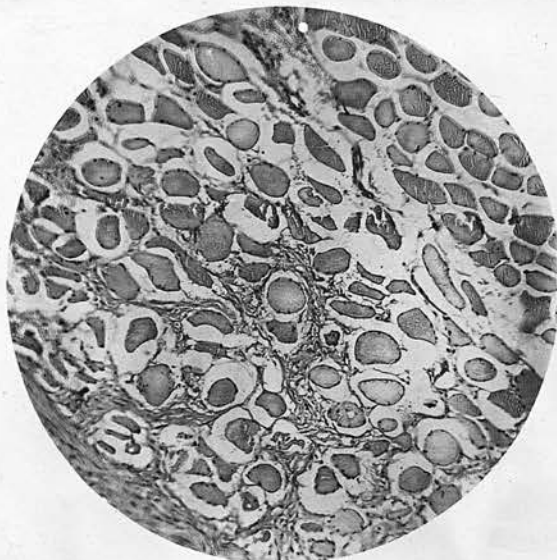


Fig.2. Microphotograph of section, Case 2, B.M,
same magnification.

Section through muscle close to growing edge
of tumour.

Note the "pseudo giant celled" appearance of
the degenerating muscle.

The interpretation which I put on these cellular masses is quite opposed to any such explanation of their origin, because in the serial sections which I have had cut we see that as the growing tumour edge invades the adjacent muscle, the muscle fibres undergo retrogressive changes only: they show hyaline degeneration, stain faintly, and their nuclei disappear (fig.2): whereas the multinucleated protoplasmic masses which lie in contact with the degenerating muscle stain well and contain abundant and well stained nuclei.

Further the view that active syncytial masses could be derived from invaded and degenerating striped muscle is opposed to all the current views as to the behaviour of injured striped muscle.

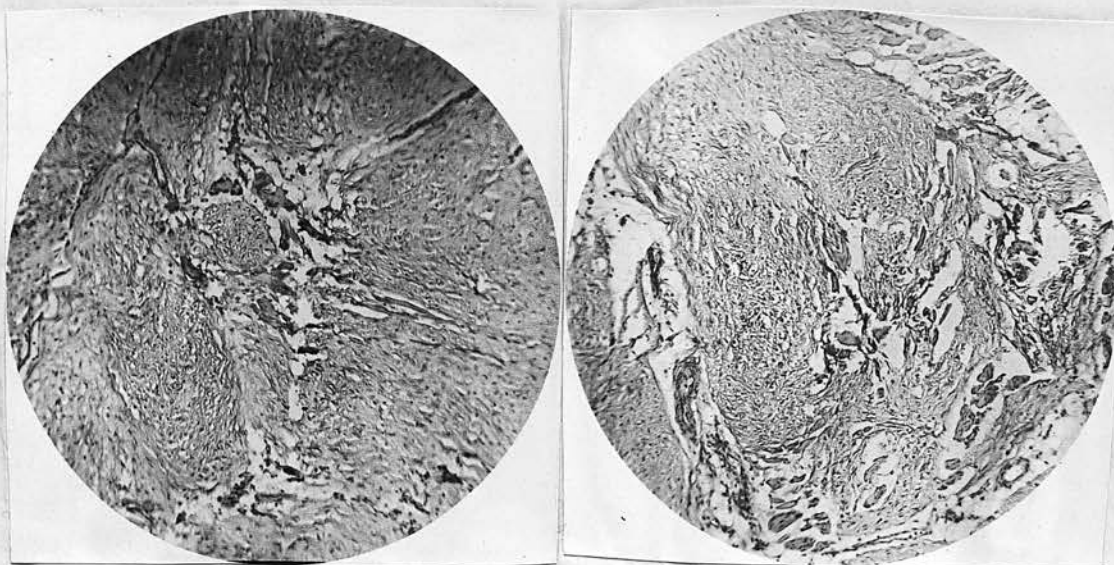
I would therefore suggest that these syncytial masses are of foetal origin and not derived from breaking up muscle fibres; and it is interesting to note with reference to their supposed sarcoplasmic staining and muscular origin, that, on similar grounds Menge in reporting two cases of Chorion-epithelioma Malignum regarded the cell masses of that tumour as sarcomatous in nature and derived from the uterine muscle, - a view which was disproved by the work of Sanger and Marchand.

I would further suggest that Desmoid tumours bear some analogy to the tumours of Chorion-epithelioma Malignum, (figs. 3; 4; and 5), - particularly to that variety of Chorion-epithelioma Malignum in which we find these characteristic tumours outside the uterus following or very rarely associated with a normal pregnancy.

We can only claim analogy with this rarer variety of Chorion-epithelioma Malignum, (a variety which constitutes 26% of recorded cases) because Desmoid tumours in common with this variety appear to almost invariably follow a normal pregnancy. This may possibly be because trauma sustained during labour supplies the necessary point of muscle injury, in which a minute haemorrhage deposits the circulating foetal epiblastic cells which initiate the growth.

Let us admit for a moment that Desmoid tumours do bear some resemblance to this particular variety of Chorion-epithelioma Malignum with regard to which Teacher and Eden in the article on Chorion-epithelioma Malignum in "Clifford Allbutt's "System of Gynaecology" write as follows :-

"The more probable explanation of these extra uterine cases /



Figs. 3 and 4. - Microphotographs of Sections of growing edge of tumour in Case 1, G.F.
(Same magnification as in Fig. 1).

- To show multinucleated protoplasmic masses in the tumour edge.

Note resemblance of those to the syncytial masses of Fig.5.

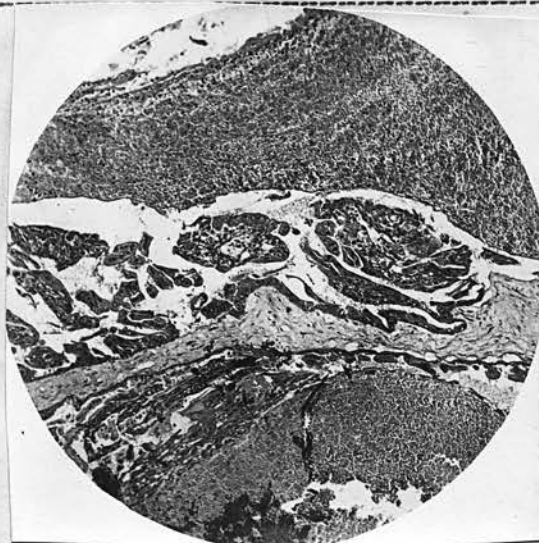


Fig. 5. - Section of Chorion-epithelioma Malignum.
(same magnification).

- To contrast with above. The haemorrhagic stroma differs markedly from the loose Fibro cellular interstitial tissue of the Desmoid.

'cases appears to us to be that they are due to detached portions of a normal placenta which assumed the characters of malignant growths in their extra-uterine seat. For it has been shown that portions of syncytium and even portions of Villi may be detached into the circulation in normal pregnancies".

So also Schmorl Zentralblatt für Gynak. 1905 makes the following statement as to the "Fate of Placental Emboli after Deportation" :-

"It is a well-established fact that primary Chorion-epithelioma Malignum occurs in the vagina, brain, and other organs and I am inclined to think with Marchand that these growths are due to the fact that deported cells from a normal placenta have the power of chorion-epitheliomatous proliferation".

The evidence of the above authorities clearly establishes the fact that foetal epithelial elements and occasionally even placental villi enter the circulation and become deposited in the maternal tissues, in which they occasionally give rise to tumour formation.

I think, therefore, we must admit that it is possible that Desmoid tumours may be originated by the deposit and growth of such deported cells, for these also are tumours which practically invariably affect women and women who have borne children: - tumours which slowly infiltrate muscle and have in their growing edge multinuclear protoplasmic masses which resemble the most characteristic cell elements of Chorion-epithelioma Malignum.

As to how this deposit actually occurs, I have suggested above that this may possibly be effected by some slight haemorrhage into muscle fibres injured by the strain and trauma of labour.

As a result of this intramuscular extravasation, cells which are specialised to invade uterine muscle are let loose in an abnormal muscular site, which reacts to their invasion.

- MORBID ANATOMY. -

The relations of Desmoid tumours to the muscle in which they /

they arise vary with their size and point of origin in the muscle. While still small they are generally found completely embedded in its fibres, which appear to terminate abruptly on the face of the tumour.

As the tumour increases in size, its growth tends to be more rapid in the direction of the muscle fibres, so that the tumour assumes an oval shape,

The larger tumours and those which have arisen near the surface of the muscle, project out from it and form firm adhesions with any fascial structures with which they may be in contact.

The tumour itself feels densely hard, in striking contrast with the muscle which embeds it.

On attempting to remove the tumour it is found to be intimately adherent to the surrounding tissues, - to "shell it out" is impossible: the tumour must be cut out from its muscular and fascial bed, and where the peritoneum, has become involved, it also must then be opened, in the great majority of cases, to permit of a complete removal.

The tumour when cut into is densely hard, creaks under the scalpel, and the cut surface is white and glistening in appearance, resembling rather closely a scirrhus tumour of the breast.

It would seem, however, as if the amount of fibrous tissue in the tumour might depend to some extent on its site: for the tumours removed from the Iliac regions are harder and more densely fibrous in nature than those of the Rectus muscle. It is possible that the tumours which arise in the groin sooner reach the limits of muscle and become adherent to, and limited by, but slightly vascular fascial structures and hence have a poorer blood supply.

An interesting point with regard to these tumours is that in no case has any associated glandular enlargement been observed.

The lymphatics of the abdominal parietes of the middle and lower abdomen drain into the Inguinal glands and this group would be affected in the large majority of these tumours: as these glands are easily palpable, it is unlikely that any enlargement would have escaped notice.

HISTOLOGY: /



HISTOLOGY: On cutting sections of the tumour we find that there is a distinct tendency for it to become more cellular in type, and looser in structure, as we pass from the central portion towards the growing edge. Sections of the centre of the tumour are like those of a typical fibroma, whereas those taken nearer the margin appear to more closely resemble the Fibro-sarcomatous type, (fig.6), till finally we reach the infiltrating margin itself, which is characterised by a still looser and more cellular structure in which we see the multinucleated cell masses and islands of degenerating muscle cut off by the advancing edge of the growth.

These variations in character of the sections from different portions are well marked and may explain the discrepancy which exists in the reports as to the pathological nature of this class of tumour and I would again emphasise the fact that a certain diagnosis of Desmoid tumour can be arrived at only by the examination of the growing edge.

The multinucleated cell masses which are here seen are of very irregular shape: they stain deeply with eosin and their nuclei - which are scattered irregularly throughout the protoplasm - stain well with chromatin stains.

No cells resembling those of Langan's layer could be recognised: but the cells of Langan's layer tend to atrophy after the 3rd month, and the deportation of the syncytial cells may have taken place at a later period of embryonic growth.

The walls of the vessels of the tumour are well-formed and the vascularity increases towards the growing edge.

The densely fibrous central portion of the Desmoid tumour differs widely from the stroma of the tumours in Chorion-epithelioma Malignum. It is very much less vascular and appears to be derived from the hypertrophied interstitial tissue of the invaded muscle, which shows a marked round celled infiltration and increased vascularity for some distance in advance of the tumour edge.

It seems, in short, as if the power which this tumour possesses of invading muscle were in large measure due to the aberrant special function of the deported foetal cells which its growing edge contains. These cells enable it to advance into, and progressively destroy, the muscle fibres, while they merely stimulate the growth of its non-specialised interstitial tissues, which hypertrophy, multiply, and later settle /

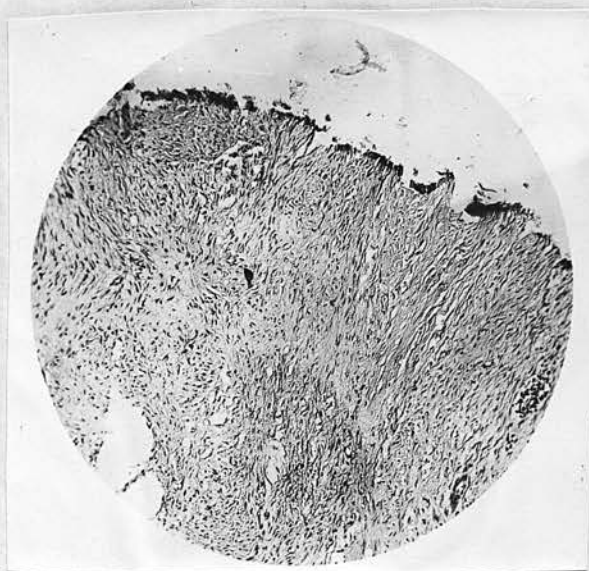


Fig.6 - Microphotograph of section cut at some distance from growing edge to show typical appearance of central portions of Desmoid Tumour.

(Same magnification as in Fig.1).

The sections of cases 3 and 4 resemble this closely and are not figured as in neither case were they cut through the edge of contact with muscle.

settle down as the growing edge leaves them and form the dense fibrous tissue which constitutes the centre of the tumour.

Finally we would draw attention to the fact that the marked differences which at first sight appear to exist between Desmoid tumours and the growths of Chorion-epithelioma Malignum are more apparent than real for the characteristic portion of each is the growing edge.

The fact that Desmoid tumours have a hard fibrous centre as contrasted with the loose haemorrhagic and necrotic core of the Deciduoma Malignum may be due to the different reaction of the tissues in which the cells initiating the growth become deposited. On the other hand the difference might be ascribed to the fact that in a Desmoid tumour we have an accidental deposit of normal circulating foetal elements whereas the great majority of Chorion-epitheliomatous growths are initiated by the deposit of abnormal foetal trophoblast which might have a purely destructive action and fail to stimulate the non-specialised portions of the invaded tissue. This latter explanation would also account for the much more serious prognosis of the Chorion-epitheliomatous growths.

- CLINICAL FEATURES. -

SIGNS and SYMPTOMS:- In the great majority of cases the presence of the tumour is discovered accidentally; - some slight blow may lead to examination and to its discovery.

The tumour is quite painless, and the ease with which it is felt varies with the depth at which it is situated in the abdominal muscles.

Such a tumour situated more superficially in the Rectus muscle feels like a hard boss in the muscle which gradually extends, becomes oval in the direction of the muscle fibres and projects till it forms a visible swelling of the abdominal wall.

It is a hard painless swelling, which does not implicate the skin and subcutaneous tissues which can be picked up and moved about freely over it, dimpled by no adhesions: a tumour, however, which is very evidently situated in the thickness of the abdominal wall and involving its muscles, because /

because it becomes fixed by their contraction. With the abdominal muscles relaxed, the tumour can be moved about with sufficient freedom to show that it is unconnected with any intra-peritoneal structure.

Desmoid tumours arising in the other common situation - the Iliac Regions, - even when comparatively superficial, are rather more difficult to diagnose, for they often render tense the fascial structures, and it is then difficult to tell whether or not they are arising from the Iliac bones. The absence of symptoms of intra-abdominal or pelvic disease may help us; while pelvic examination and an X Ray photograph will show that the bones themselves are uninvolved.

The greatest difficulty, however, is presented in the diagnosis of those tumours, the main bulk of which is situated deep to the Rectus or Oblique Muscles. I would say that it is practically impossible to arrive at a correct diagnosis - given a stout or muscular patient with a deep seated tumour. For the great difficulty lies in differentiating between such a tumour and an intra-peritoneal tumour of any of the subjacent viscera, eg. Malignant mass in omentum, Omental Hernia through back of Rectus sheath.

Where we have from our physical examination come to the conclusion that the tumour is undoubtedly situated in the abdominal wall, we have still to exclude it from other tumours solid and cystic which may occupy a similar site, - From Subcutaneous Nodule; from Myomata arising from the Round Ligaments; from Haematomata due to trauma; from Tuberculous Abscess; from Sarcomata of muscle, periosteum, or bone.

The chief points which would aid us in forming a diagnosis of Desmoid tumour are as follows :-

- The patient is a robust woman who has borne one or more children. She now presents herself with a hard, painless, fairly rapidly growing tumour situated either in the Rectus Muscle or in the abdominal wall of either Iliac Region. The tumour is partly or wholly within the muscle and is fixed on contraction of the muscle. The tumour can be moved about over the contents of the abdomen. There are no symptoms of intra-abdominal disease.

COURSE and PROGNOSIS:- The rate of growth of Desmoid tumours /

tumours is fairly rapid but varies widely in different recorded cases. An associated pregnancy appears to stimulate growth, and haemorrhage into the tumour may be determined by trauma incidental to the labour. Spontaneous disappearance has never been recorded.

Where the tumour is left to itself it may attain a large size, - eg. 6 kilos in tumour of 10 years' duration (Gross & Sencert) and, as in this case, these large tumours show a tendency to become pedunculated and to bulge either externally or subperitoneally.

It is possible that a tumour in the former situation might finally through ulceration of its coverings become septic and gangrenous: while the subperitoneal tumours of large size would cause intra-abdominal pressure symptoms.

Death as a direct result of the tumour must be very rare as they have no tendency to form metastases, - nor do they ever spread to intra-peritoneal structures.

PROGNOSIS as regards OPERATION:-

Pfeiffer writes as follows with regard to the results of operation :- "Since the introduction of asepsis the immediate results of operation are relatively favourable and we presume that antiseptic or aseptic operation methods were used in the 285 cases recorded in the last 14 years".

In 119 of these 285 -(40.8%) the peritoneal cavity was opened during operation and immediate mortality was 3.5%.

In 142 (49.8%) tumour extirpated without opening peritoneum, with immediate mortality of1.05%.

(Details as to remaining 24 cases were insufficient).

Pfeiffer gives the following data as to after history of 107 cases (22 men; 85 women).

Of 22 men..... 15 recurred = 68.1%
of these 11 died= 50% mortality.

while /

while 18 of the 85 women showed recurrence = 21.2%
and of those 6 died mortality = 7%.

Of these 33 cases in which recurrence took place after operation :-

In 22.....it took place within the first year.

" 5.....during the second year.

" 6.....at some time after the end of second year.

No case of dissemination or glandular involvement was recorded.

We see from the above figures that where recurrence after operation does occur, it is always local and shows itself with very great frequency during the first year. The figures on this point are so remarkable that I am led to conclude that these are not instances of recurrence in the true sense of the term, but are rather due to the continued growth of fragments of a tumour which has been incompletely removed.

The above figures tend to support the conclusion which we formed, - i.e. that the growing edge alone is characteristic and endows the tumour with the power of progressive infiltration of muscle. A thorough removal of this growing edge is therefore essential if we would avoid continued growth and reformation of the tumour.

The widely different prognosis of so-called "Desmoid" tumours in the male sex can only be explained by the presumption that they are of a different pathological nature.

The immediate operation mortality is due to sepsis and occasionally to haemorrhage, and, where, as in several recorded cases, death occurred after re-operation for so-called recurrence, in these cases also it was due to defective operative technic and not to the tumour per se.

The immediate operation mortality of the 35 cases collected for this thesis appears to have been nil and recurrence is stated to have taken place in only 3 instances.

[illegible]